```
Trying 31060000009999...Open

DIALOG INFORMATION SERVICES
PLEASE LOGON:
 ******** HHHHHHHH SSSSSSSS? ### Status: Signing onto Dialog ********
ENTER PASSWORD:
 ******** HHHHHHHH SSSSSSSS?  ********
### Status: Login successfulWelcome to DIALOG

Dialog level 05.06.01D

Last logoff:  12oct05 12:56:55
Logon file405 13oct05 12:36:50
           *** ANNOUNCEMENT ***
                    ***
--UPDATED: Important Notice to Freelance Authors--
See HELP FREELANCE for more information
                    ***
NEW FILES RELEASED
***Computer and Information Systems Abstracts (File 56)
***Electronics and Communications Abstracts (File 57)
***Solid State and Superconductivity Abstracts (File 68)
***ANTE: Abstracts in New Technologies (File 60)
                    ***
RESUMED UPDATING
***ERIC (File 1)
                    ***
Chemical Structure Searching now available in Prous Science Drug
Data Report (F452), Prous Science Drugs of the Future (F453),
IMS R&D Focus (F445/955), Pharmaprojects (F128/928), Beilstein
Facts (F390), and Derwent Chemistry Resource (F355).
                    ***
     >>> Enter BEGIN HOMEBASE for Dialog Announcements <<<
     >>>    of new databases, price changes, etc.      <<<
                    ****
* * *
SYSTEM:HOME
Cost is in DialUnits
Menu System II: D2 version 1.7.9 term=ASCII
                      *** DIALOG HOMEBASE(SM) Main Menu ***

 Information:
  1.  Announcements (new files, reloads, etc.)
  2.  Database, Rates, & Command Descriptions
  3.  Help in Choosing Databases for Your Topic
  4.  Customer Services (telephone assistance, training, seminars, etc.)
  5.  Product Descriptions

 Connections:
  6.  DIALOG(R) Document Delivery
  7.  Data Star(R)

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      /H = Help           /L = Logoff          /NOMENU = Command Mode


Enter an option number to view information or to connect to an online
 service.  Enter a BEGIN command plus a file number to search a database
(e.g., B1 for ERIC).
?
```

```
Terminal set to DLINK
                    *** DIALOG HOMEBASE(SM) Main Menu ***

 Information:
  1.  Announcements (new files, reloads, etc.)
  2.  Database, Rates, & Command Descriptions
  3.  Help in Choosing Databases for Your Topic
  4.  Customer Services (telephone assistance, training, seminars, etc.)
  5.  Product Descriptions

 Connections:
  6.  DIALOG(R) Document Delivery
  7.  Data Star(R)

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      /H = Help          /L = Logoff          /NOMENU = Command Mode


Enter an option number to view information or to connect to an online
 service.  Enter a BEGIN command plus a file number to search a database
(e.g., B1 for ERIC).
? b 71, 73, 144

       13oct05 12:37:01 User276793 Session D25.1
            $0.00    0.211 DialUnits FileHomeBase
     $0.00  Estimated cost FileHomeBase
     $0.05  TELNET
     $0.05  Estimated cost this search
     $0.05  Estimated total session cost   0.211 DialUnits


SYSTEM:OS  - DIALOG OneSearch
  File  71:ELSEVIER BIOBASE 1994-2005/Oct W1
        (c) 2005  Elsevier Science B.V.
  File  73:EMBASE 1974-2005/Oct 13
        (c) 2005 Elsevier Science B.V.
  File 144:Pascal 1973-2005/Oct W1
        (c) 2005 INIST/CNRS

      Set  Items  Description
      ---  -----  -----------
? s liver and (gene (w) therapy) and (ductal (w) delivery)
          824819  LIVER
         1677191  GENE
         2935982  THERAPY
           54631  GENE(W)THERAPY
           24872  DUCTAL
          271737  DELIVERY
               0  DUCTAL(W)DELIVERY
      S1       0  LIVER AND (GENE (W) THERAPY) AND (DUCTAL (W) DELIVERY)
? s liver and (gene (w) therapy)
          824819  LIVER
         1677191  GENE
         2935982  THERAPY
           54631  GENE(W)THERAPY
      S2    5026  LIVER AND (GENE (W) THERAPY)
? rd
>>>A maximum of 5000 items can be processed.
? s s2 not py>=1996
            5026  S2
```

```
        11896952  PY>=1996
      S3     506  S2 NOT PY>=1996
? rd
...examined 50 records  (50)
...examined 50 records  (100)
...examined 50 records  (150)
...examined 50 records  (200)
...examined 50 records  (250)
...examined 50 records  (300)
...examined 50 records  (350)
...examined 50 records  (400)
>>>Record 144:12440130 ignored; incomplete bibliographic data, not retained
 in RD set
...examined 50 records  (450)
>>>Record 144:11684135 ignored; incomplete bibliographic data, not retained
 in RD set
...examined 50 records  (500)
...completed examining records
      S4     375  RD (unique items)
? t/full/1 from each
```

4/9/1 (Item 1 from file: 71)

00357253 96008682

The sparse fur mouse as a model for gene therapy in ornithine carbamoyltransferase deficiency

Batshaw M.L.; Yudkoff M.; McLaughlin B.A.; Gorry E.; Anegawa N.J.; Smith I.A.S.; Hyman S.L.; Robinson M.B.
ADDRESS: M.L. Batshaw, 3405 Civic Center Boulevard, Philadelphia, PA 19104-4399, United States

Journal: Gene Therapy, 2/10 (743-749), 1995, United Kingdom
PUBLICATION DATE: 19950000
CODEN: GETHE
ISSN: 0969-7128
DOCUMENT TYPE: Article
LANGUAGES: English SUMMARY LANGUAGES: English

The sparse fur (spf/Y) mouse was evaluated as a model for studying **gene therapy** in ornithine carbamoyl-transferase deficiency (OCTD), the most common inborn error of urea synthesis. Previous studies have defined a number of biochemical characteristics of this animal model that are analogous to the human disease: OCTD in **liver** , elevated ammonium and glutamine, low citrulline and arginine in plasma, elevated urinary orotic acid excretion, neurochemical alterations and responsiveness to alternative pathway therapy. In this study, metabolic flux, survival, behavior and learning of these animals were examined in preparation for a trial of **gene therapy** . We found that, as has been previously reported. OCT activity in **liver** ranged from 10 to 20% of control. Yet, stable isotope studies using sup 1sup 5N ammonium chloride to follow ureagenesis in vivo showed 55% of normal urea synthetic capacity. This suggests that partial correction with **gene therapy** may be sufficient to normalize urea synthesis. Although it has been suggested that **liver** OCTD and its consequent metabolic effects normalize without treatment by adulthood in the spf/Y mouse, we did not find this to be the case. We documented that the spf/Y mouse had a markedly decreased lifespan (< 10% of normal) and remained runted throughout life. In terms of behavior, the spf/Y mice had evidence of decreased learning in a passive avoidance task that was not attributable to alterations in activity. These clearly definable metabolic and behavioral abnormalities

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